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Quality of Care and Outcomes Assessment

LONG-TERM FOLLOW-UP IN MARFAN PATIENTS AFTER SUCCESSFUL SURGERY FOR ACUTE TYPE A AORTIC DISSECTION. THE IRAD EXPERIENCE

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Backgroud: Life expectancy of patients (pts) with Marfan Syndrome (MS) is shorter than that of the general population. The long-term outcome after surgery for aortic dissection (AD) is less clear. The large patient cohort in IRAD facilitates the investigation of long-term outcomes in surgically managed acute type A AD pts with MS compared to those without the disease.

Methods: Our study evaluated 775 surgically-managed Type A AD pts enrolled in IRAD between 1996 and 2011, with follow-up available at 1, 2 and/or 3 years post-discharge. The pts were divided into groups based on presence (n=36, 4%) or absence of MS. Categorical variables were analyzed using chi-square test and Fischer's exact test where applicable. Continuous variables were analyzed using student's t-test for normally distributed data, and test of medians for non-normally distributed data.

Results: The mean age was significantly lower for MS pts (34.8 ±10.3 years vs 60.0 ± 12.4 years [p <0.001]). They also had more aortic insufficiency at presentation (68.8% vs 39.5% [p=0.001]). More MS pts were normotensive on presentation (64.7% vs 44.1% [p=0.018]). Significantly more concomitant procedures were performed in the MS group (60% compared to 36% [p=0.004]). Their aortic root dimension was larger (5.8 cm compared to 4.2 [p<0.001]). Extension of dissection, defined as post-operative involvement of additional arteries that resulted as a sequelae of the AD, was higher in the MS (9.1%) compared to the non-MS group (1.5%) (p=0.021). Systolic blood pressure was lower at one year (119.2 ± 12.4 vs 130.3 ± 21.5 [p=0.036]) and at three years (114±5 vs 131±17 [p=0.026]) in MS pts. Also at three years the total aortic diameter was significantly increased in 66.7% of MS pts compared to 21.3% in the non-MS group (p=0.024). Kaplan-Meyer survival curve showed no significant difference in survival between the two groups from discharge to three year follow-up.

Conclusion: Our data show that MS pts after surgery for type A AD are more likely to have extension of dissection and increased aortic diameters on follow-up, despite lower systolic blood pressure and similar mortality at three years post-op. This suggests closer follow-up is important for pts with MS.